Title
Intracranial Hypertension in a Middle-aged Women Secondary to Levonorgestrel Intrauterine Device ("Mirena")

Abstract
Idiopathic intracranial hypertension (IIH) is characterized by elevated intracranial pressure, normal CSF composition, and normal neuro-imaging. Intracranial hypertension and its associated signs and symptoms have been observed in women implanted with the “Mirena” IUD.

Case History

Patient demographics: 52-year-old Caucasian female

Chief complaint
The patient reported: “I see black spots in my eyes which synchronize with my heartbeat, and ringing in my ears when I stand up or sit down.” She also complained of blurred vision with her current glasses in both eyes.

Ocular history
Chronic recurrent idiopathic intracranial hypertension, Irvine Gass syndrome, pseudophakia, dry eyes, lagophthalmos, hypertensive retinopathy

Medical history
Headaches, hearing loss, hypertension, Bell’s palsy, diabetes mellitus, sleep apnea, metrorrhagia, asthma, anxiety, posttraumatic stress disorder, agoraphobia

Medications
Systemic: Lisinopril 10mg tab, insulin, metformin hcl 1000mg tab, albuterol sulfate inhaler, aripiprazole 2mg tab, fluoxetine hcl 20mg capsule

Ocular: Acular 0.5% oph solution, Pred Forte 1% oph suspension

Surgeries
Insertion of intrauterine device (Mirena)
Cataract extraction OU

Allergies
Diamox, epinephrine, risperidone, prazosin, atorvastatin

Family ocular or medical history
Unremarkable

Pertinent findings

Clinical findings
- Blood pressure: 144/83 (ranges 140s systolic and 85 diastolic)
- BCVA: 20/40 OD and 20/50 OS at distance
- Confrontation visual fields: Full to finger count OD and OS
- Color testing (Ishihara plates): 13/14 OD, 13/14 OS
- Pupil testing: pupils round, equal, and responsive to light; 0.3 APD OD only
- Extraocular motility: Smooth full range of motion with no pain or diplopia
- Anterior segment: 1+ scattered punctate epithelial erosions OU, pterygium nasal OS
- Intraocular pressure (via Goldmann tonometry): 15mmHg OD and 14mmHg OS
- Posterior segment: Grade 3 disc edema OU, macular edema OU, mild hypertensive retinopathy OU
**External/Physical findings**

Obesity, facial asymmetry secondary to Bell’s palsy

**Laboratory studies**

- Labs were normal with the exception of elevated glucose, elevated cholesterol levels, decreased vitamin D

**Radiology studies**

- MRI: revealed no masses or vascular abnormalities; however edema of both optic nerve sheaths and hyperintensity at the CN VII nucleus were noted
- MRV/MRA: no signs of venous sinus thrombosis
- Lumbar puncture: elevated opening pressures (25cm)

**Others**

- Color fundus photos: bilateral swollen and hyperemic discs with blurred margins and obscured vessels
- Optical coherence tomography (macula): cystoid macular edema OU
- Optical coherence tomography (optic nerve): thickened RNFL layer OU
- Goldmann visual field (GVF): marked constriction of all isopters with enlarged blind spots OU
- Intravenous fluorescein angiography (IFVA)
  - Optic nerve: hyper-fluorescence of the optic nerve with no auto-fluorescence prior to injection OU
  - Macula: early hyper-fluorescence of the macula OU

**Differential diagnosis**

**Primary/leading diagnosis**

Primary: Idiopathic intracranial hypertension

**Others**

1. **Pseudopapilledema (optic nerve head drusen, tilted disc, myelinated nerve fibers)**
   - **Presentation:** vessels overlying disc are not obscured, optic discs are not hyperemic, and spontaneous venous pulsations are present.
   - Diagnosis was ruled out based on absence of auto-fluorescence on IVFA, vessels overlying disc were obscured, absence of tilted disc and myelinated nerve fibers on fundus exam.

2. **Papillitis (optic neuritis)**
   - **Presentation:** swollen discs, pain on eye movements, afferent pupillary defect, decreased vision in one eye, and decreased color vision, common in children and young adults
   - Diagnosis was ruled out based on normal MRI brain imaging, full EOMs without pain, normal color vision, and age.

3. **Central retinal vein occlusion**
   - **Presentation:** hemorrhages extending far beyond peripapillary area, dilated and tortuous veins
   - Diagnosis ruled out based on absence of retinal hemorrhages and normal vessel caliber.

4. **Hypertensive retinopathy**
   - **Presentation:** extremely high blood pressure with signs of severe retinopathy (ie. Hemorrhages, cotton wool spots, and exudates).
   - Diagnosis was ruled out because blood pressure was not in malignant range in office and fundus showed only mild retinopathy.
5. Leber hereditary optic neuropathy
   - **Presentation:** rapid vision loss; usually occurs in men in the second and third decade with a family ocular history of this condition.
   - Diagnosis was ruled out based on gender, unremarkable family ocular history, and absence of rapid vision loss.

6. Inflammatory optic neuropathy (demyelinating, sarcoidosis, infectious)
   - Diagnoses ruled out based on normal lab results and normal chest X-ray

7. Ischemic or vascular optic neuropathy (AAION or NAAION)
   - **Presentation:** Pale, swollen disc, with flame-shaped hemorrhages, APD, and visual loss.
   - Diagnosis was ruled out based on normal blood tests, mild attenuation of the retinal vessels, age, and denial of signs/symptoms of GCA.

8. Carotid-cavernous fistula
   - Diagnosis was ruled out based on normal MRI imaging.

9. Compressive optic neuropathy
   - Diagnosis was ruled out based on normal MRI imaging.

**Diagnosis and discussion**

Idiopathic intracranial hypertension (IIH) is characterized by increased intracranial pressure with the following criteria: symptoms and signs of increased intracranial hypertension or papilledema, elevated CSF opening pressure during lumbar puncture, normal CSF composition, and normal MRI imaging.¹

Patients have symptoms of headaches, nausea, pulsatile tinnitus, papilledema, transient visual loss often precipitated after rising from a lying or sitting position, and diplopia from sixth nerve palsy. Most patients are female of childbearing age and at least 20% over their ideal body weight.²

While the etiology is unknown, suggested mechanisms include increased production and/or reduced absorption of CSF, sustained elevated intracranial venous pressure, and possible increased cerebral blood volume. Associated factors of IIH are obesity and pregnancy. Possible causative factors include oral contraceptives, tetracycline (and semisynthetic derivatives), vitamin A, cyclosporine, amiodarone, sulfa antibiotics, lithium, systemic steroid intake and/or withdrawal.² Other forms of contraceptive delivery, such as intrauterine devices, have also been linked to idiopathic intracranial hypertension.⁴

**Treatment/Management**

The goals of managing IIH include relieving headaches, diplopia, and preserving vision. Lumbar puncture is usually the first step of treatment as it immediately decreases intracranial pressure. Other treatments include weight loss if overweight, Acetazolamide (Diamox), and discontinuation of any causative medications.² Surgical treatments include CSF shunting procedures, optic nerve sheath fenestration, and endovascular venous stenting. Surgeries are reserved for when conservative treatments are unsuccessful and/or headaches are severe and intractable.³

**Conclusion**

Oral contraceptive medications are one of the most commonly prescribed classes of medications used by young women, however there are a considerable number of women who prefer other forms of contraceptive delivery such as intrauterine devices.
Mirena (IUD) is an intrauterine device in the class of intrauterine devices called levonorgestrel intrauterine systems. It is placed in the uterus and slowly releases the birth control hormone progestin into the body. It acts as long-term contraception for up to five years. It would be expected to be safer than oral contraceptives due to its relatively low dose of hormone release. The presence of the progestin levonorgestrel, which is the synthetic birth control hormone used in the Mirena, are reported to increase the risk of idiopathic intracranial hypertension.4

There are many risk factors that have been associated with IIH. Recognizing and understanding the risk factors that truly contribute to intracranial hypertension is important in both diagnosing and understanding the pathophysiology of the disease. Fortunately, this condition can be managed by treating the cause of the condition to preserve visual function. Most patients will have complete resolution of symptoms without persistent deficits if they are compliant with regular follow-up visits and treatments.

References


